VALVULOTOMY FOR THE RELIEF OF CONGENITAL VALVULAR PULMONIC STENOSIS WITH INTACT VENTRICULAR SEPTUM

REPORT OF NINETEEN OPERATIONS BY THE BROCK METHOD*

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The first attempt to treat valvular stenosis by surgery was made by Doyen in 1913 and is described by J. Dumont.[1] The patient was believed to have valvular pulmonic stenosis, and the treatment consisted of an attempt to relieve the stenosis by dividing the lesion with a small tenotomy knife which was introduced through the wall of the right ventricle. The patient survived only a few hours, and necropsy demonstrated a subvalvular rather than a valvular stenosis. No further attempts were made to treat valvular pulmonic stenosis by a direct attack on the valve itself until recently; in the intervening time more attention has been focused on the surgical treatment of mitral stenosis. The results in the 1920-30 period were rather discouraging, whereas those in the past several years have shown considerable improvement. Recent progress in the treatment of valvular pulmonic stenosis has paralleled the advance in the treatment of mitral stenosis. Russell Brock[2] reported (June 12, 1948) the results of operations on three patients in whom the stenotic pulmonary valve had been divided during the preceding four months. Holmes Sellors[3] reported (June 26, 1948) a single case in which the pulmonary valve had been divided successfully almost eight months previously. In his last published report in August, 1949, Mr. Brock[4] described the ten patients in whom pulmonary valvulotomy had been attempted. The result was excellent in two of these; in two it was considered to be moderately satisfactory, and in one the operation was too recent to permit evaluation of the result. Three of the five deaths were due to cardiac failure which occurred shortly after the chest was opened. It will be brought out later that Mr. Brock's recent experience while visiting in this country was much more favorable.

In congenital stenosis of the pulmonary valve there is fusion of the three semilunar cusps to form a domelike structure with a small central perforation (Fig. 1). During systole this diaphragm projects into the pulmonary artery and a small but powerful jet of blood is forced through the valve. It is probably this jet effect which is responsible for the poststenotic dilatation

* Aided by a Grant from the Robert Garrett Fund for the Surgical Treatment of Children and by the Gift of Mr. Hal Price Headley of Lexington, Kentucky. Read before the American Surgical Association, Colorado Springs, Colorado, April 20, 1950.

which is usually seen in this condition. In some patients the pulmonary artery is dilated to several times its normal size, assuming almost aneurysmal proportions. The pressure within the artery is, however, low rather than high. Proportional to the severity of the stenosis and to the age of the patient, there is right ventricular hypertension and secondary right ventricular and right auricular hypertrophy and dilatation.

It has been customary to refer to valvular pulmonic stenosis occurring without an interventricular septal defect as "pure" pulmonic stenosis. It should, however, be remembered that in about 70 per cent of these patients the foramen ovale has been held open by the elevated right auricular pressure; a right to left shunt is thus produced which may involve more than half of the venous return to the right auricle. This fact alters the hemodynamics of

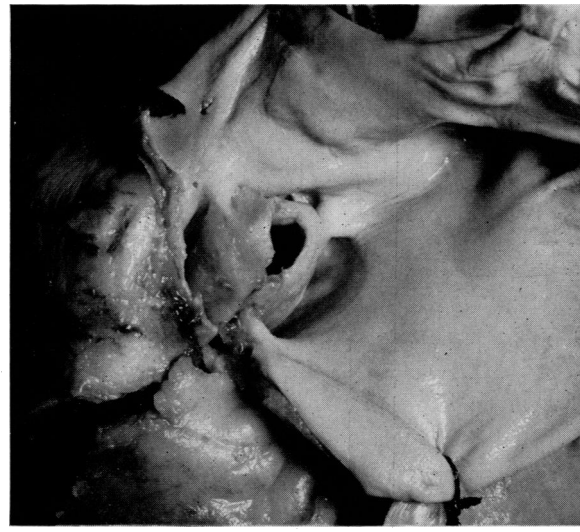


FIG. 1.—This shows the domelike structure formed by the fusion of the three cusps of the pulmonary valve. Since this valve has been cut with a valvulotome, the degree of stenosis appears less than was actually the case.

the condition in that an escape valve has been provided which to some degree decompresses the right auricle and may delay the onset of right-sided heart failure. It also introduces the factor of cyanosis and increases the left ventricular output. It is interesting to note that in one of our patients (A. V.) the mother observed that as his cyanosis increased his exercise tolerance improved. We attributed this improvement to an increase in the right to left shunt through the foramen ovale.

Valvular pulmonic stenosis in association with a patent foramen ovale can easily be confused with a tetralogy of Fallot, as in three of the patients reported herein. It is important from the viewpoints of both diagnosis and surgical therapy that one should clearly understand the anatomical and

mechanical differences between these two conditions (Figs. 2 and 3). In the majority of cases of tetralogy the stenosis is in the pulmonary conus of the right ventricle, the infundibulum, rather than in the valve itself. As pointed out by Brock, a direct attack on the stenotic infundibular region may be feasible in some of the patients with the tetralogy of Fallot, particularly those with a large infundibular chamber between the region of stenosis and the valve. This approach, however, is a much more difficult and dangerous one than the attack on the stenotic pulmonary valve. Another important

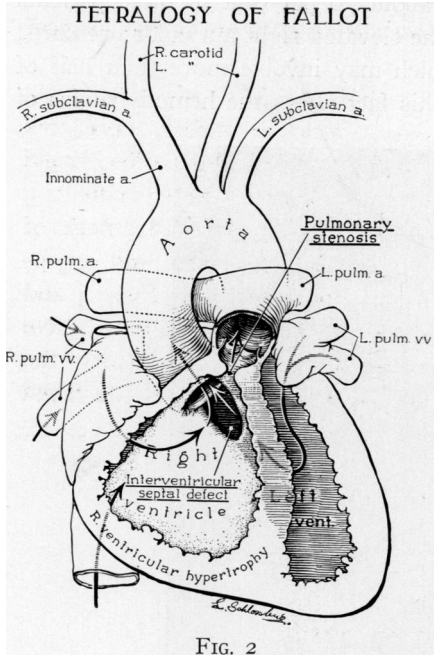


FIG. 2

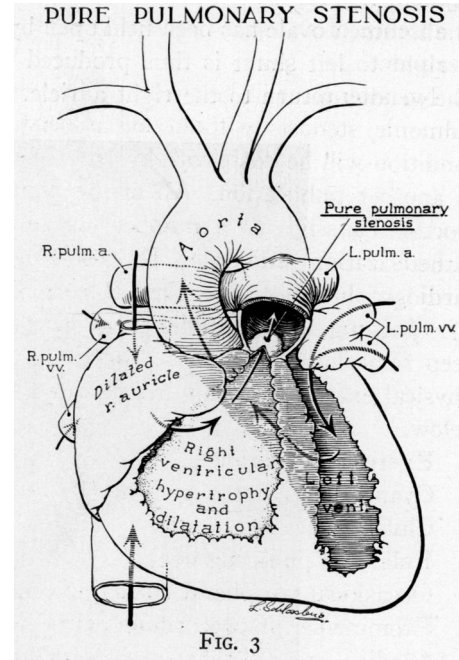


FIG. 3

FIG. 2.—Diagram of tetralogy of Fallot—note stenosis of infundibulum with the infundibular chamber lying between the point of stenosis and the essentially normal pulmonary valve. There is a high interventricular septal defect with overriding of the aorta.

FIG. 3.—Diagram of "pure" pulmonic stenosis—note fusion of valvular cusps to form diaphragmlike structure with small central perforation. The interventricular septum is intact and the main pulmonary artery beyond the valve is dilated. There is a greater degree of hypertrophy of the right ventricle and right auricle than is seen in the tetralogy of Fallot. Patency of the foramen ovale is not indicated, but is usually present.

difference between the two conditions lies in the fact that in the tetralogy of Fallot there is an interventricular septal defect with overriding of the aorta. This permits the aorta to receive blood from the right ventricle as well as the left, and constitutes an escape valve which protects the right ventricle from an excessive degree of hypertension. In such a situation the mixed blood of the left ventricle can be shunted to the lungs by an anastomosis between a systemic and a pulmonary artery, with great improvement in arterial oxygen content and relatively little danger of right-sided heart failure. How-

ever, the patient with valvular pulmonic stenosis in association with a patent foramen ovale is in a far more precarious state of balance as regards his pressure relationships. Whereas such a shunt will improve his peripheral arterial oxygen saturation, it will also increase the pulmonary venous return to the left auricle and diminish the shunt through the foramen ovale which has been decompressing the right side of the heart. Such a patient can be expected to have severe right-sided heart failure shortly after operation. It is apparent, therefore, that when pulmonic stenosis occurs without an interventricular septal defect a direct attack on the stenotic area is necessary. The presence or absence of a patent foramen ovale does not alter the nature of the surgical treatment.

No attempt will be made to consider in detail the diagnosis of valvular pulmonic stenosis without an associated interventricular defect, since this condition will be dealt with by Dr. Helen Taussig and Dr. Mary Allen Engle in another publication. All of the patients reported herein were studied by Doctor Taussig. In a number of the cases we have had the advantage of catheterization studies by Dr. Richard Bing and his associates and angiocardiographic studies by Drs. Robert Sloan and Robert Cooley (Figs. 4 and 5). Judging by the findings at operation, the preoperative diagnoses have been remarkably accurate. The more important features of the history, the physical examination, and the findings by special methods of study are listed below.

Exercise intolerance.

Cyanosis if shunt is present (70 per cent), usually late onset.

Clubbing of digits.

Enlarged, pulsating liver.

Occasional peripheral edema and ascites.

Prominence of precordium with precordial heave.

Cardiac enlargement, predominantly right-sided.

Systolic murmur and thrill.

Dilatation of main pulmonary artery visible on roentgen-ray examination.

Absence of vigorous pulsations in lungs on fluoroscopy.

Polycythemia depending on degree of unsaturation.

Albuminuria.

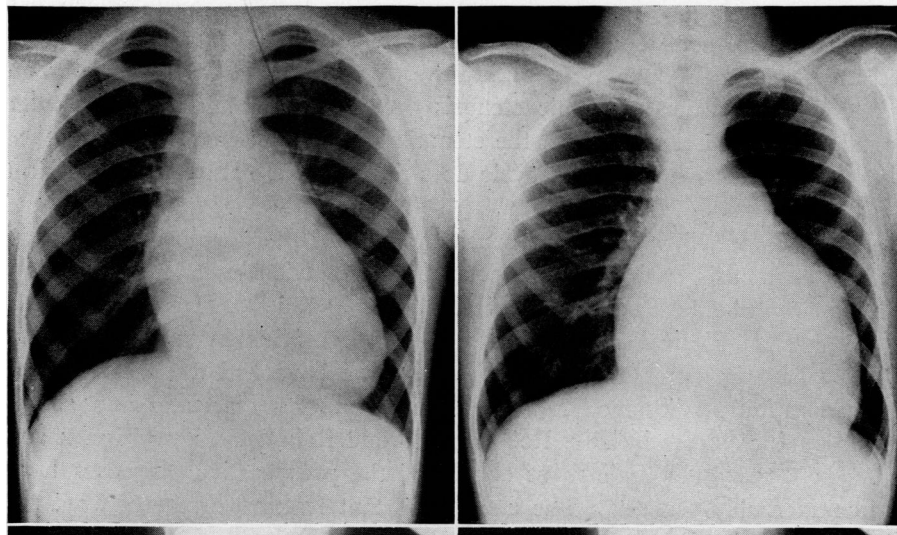
Prolonged circulation time, variable, depending on interauricular septal defect.

Electrocardiogram: right axis deviation and right ventricular hypertrophy early, right bundle branch block late.

Angiocardiography: slow passage of dye through heart unless there is large interauricular septal defect, large right ventricle, poststenotic dilatation of pulmonary artery with lingering of dye.

Catheterization: high pressure in right ventricle, low or normal pressure in pulmonary artery, diminished pulmonary flow, arterial oxygen unsaturation, depending on interauricular septal defect.

FIG. 4



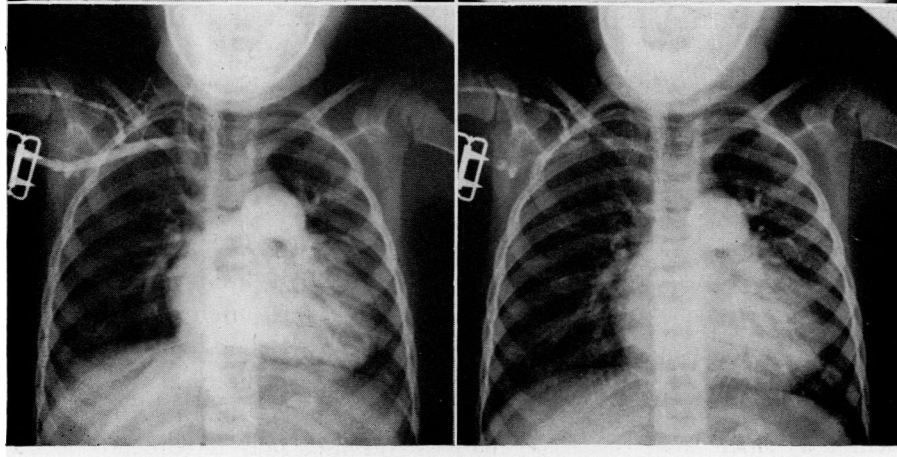


FIG. 5

FIG. 4.—Chest roentgenograms of two patients (J. T. and C. B.) discussed in text—note cardiac enlargement and prominence in region of main pulmonary artery.

FIG. 5.—Angiocardiograms of patient (E. F.) discussed in text. (A) This was taken two seconds following injection of the opaque medium and shows the marked dilatation of the main pulmonary artery. (B) This was taken two seconds later and shows the opaque medium to be lingering in the pulmonary artery and still no opacification of the aorta.

It is not necessary to employ all of the special methods of study in all cases, since the correct diagnosis may become apparent from the usual clinical and radiologic examinations.

OPERATIVE PROCEDURE

The operation is that which has been described by Mr. Brock of Guy's Hospital, London, and in fact he performed the operation on seven of the patients described in this report while he was serving as an Exchange Professor in The Johns Hopkins Medical School and Hospital. Slight variations which we have made in Mr. Brock's technic are without significance.

If the patient presents evidence of heart failure, digitalis is administered preoperatively. If cardiac irregularity is found on electrocardiographic studies, quinidine is given. On the evening prior to operation, the intramuscular administration of penicillin is begun. The pre-anesthetic medication includes the giving of morphine and atropine. Blood or plasma, depending upon the degree of polycythemia, is administered continuously during the operation through a cannula inserted into a vein in the ankle. At the time the pleural cavity is entered a peripheral vasoconstrictor with a prolonged duration of action (Drinalfa in these cases) is injected intramuscularly. When the pericardium is opened procaine 1 per cent in a dosage of 2 mg. per Kg. of body weight is given intravenously and is repeated if an arrhythmia of ventricular origin develops. At the same time 5 to 10 cc. of 4 per cent procaine are instilled into the pericardial cavity and allowed to remain for about five minutes while further dissection of the pericardium is carried out. The usual cardiac stimulants, as well as an electrical defibrillator, constitute part of the routine equipment.

The anesthesia in these cases has been very capably administered by Miss Olive Berger and her staff. It has consisted of cyclopropane for the induction, followed by ether. An endotracheal tube is inserted and respirations are manually assisted throughout the entire operation. A special effort is made to keep the lung on the operated side as well inflated as is consistent with adequate exposure of the field. Occasionally the anesthesia is supplemented by small doses of curare, but this was used only infrequently in the more recent cases. The plane of anesthesia is light and the patient is usually awake at the end of the operation.

For the operation the patient is placed on his back and a left anterolateral approach is used. The incision extends from the left margin of the sternum to the midaxillary line and the pleural cavity is entered through the third intercostal space. The incision in the pleura and muscle extends further posteriorly than the skin incision. In most instances the fourth costal cartilage is removed and the internal mammary blood vessels are ligated and divided. In most of these patients the right ventricle is considerably enlarged, and this fact makes its exposure easier from the left than would otherwise be the case. A small opening is made in the pericardium just anterior to the phrenic

nerve, and after some of the fluid has been allowed to escape, procaine is injected and is left in the cavity for several minutes. During this time the fatty tissue and thymus overlying the left side of the pericardium are dissected towards the midline. A long longitudinal incision is made in the pericardium just anterior to the phrenic nerve, and by means of two transverse incisions near the ends of the longitudinal incision a flap of pericardium with its base near the midline of the sternum is created. It is possible to dislocate the heart somewhat to the left by traction on this flap of pericardium. Procaine, 4 per cent, is applied topically to the surface of the right ventricle.

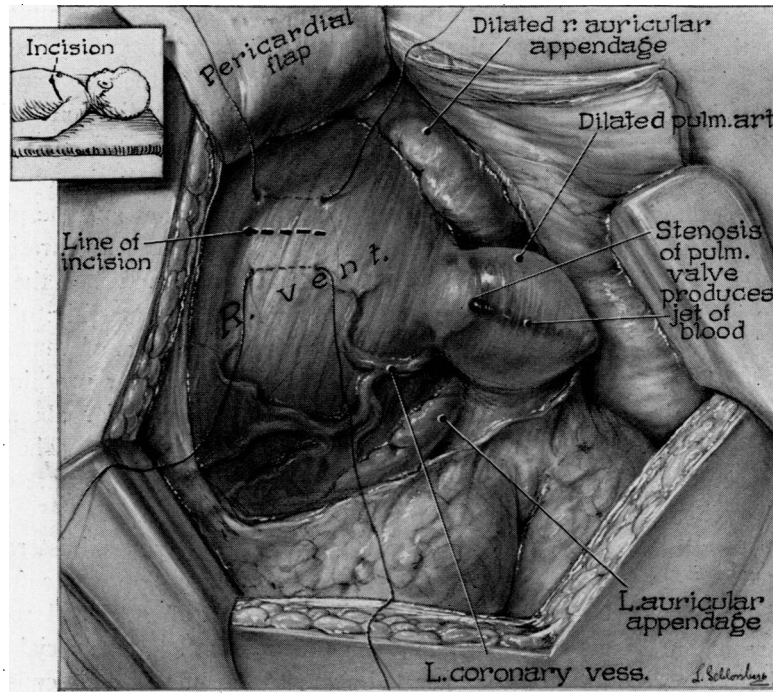


FIG. 6.—Exposure of operative field—note relation of incision to the region of the pulmonary valve and the anterior descending branch of the left coronary artery. The pulmonary artery has been made transparent to demonstrate the domelike valve and the jet of blood flowing through the small orifice.

After the pericardium has been widely opened the pulmonary artery and right ventricle and other structures are inspected and palpated. The length of time available for this examination varies; in some instances the pulse becomes poor and the blood pressure falls after the heart is exposed and one must proceed rapidly with valvulotomy. A brief examination will usually enable one to substantiate the diagnosis. The pulmonary artery in valvular stenosis is narrow at its beginning and just distally in most patients becomes a large thin-walled dilated vessel. Palpation of the first part of the dilated

pulmonary artery reveals a thrill of high-pitched frequency. The jet of blood which has passed through the stenotic valve is distinctly felt. In addition, one can usually feel by light palpation the convex surface of the dome-like shape of the stenotic valve as it is thrust forward with each systole. On palpating the region of the base of the right ventricle one does not feel the thrill that is palpable over the pulmonary artery. At the base of the pulmonary artery the bulbous distention of the sinuses of Valsalva usually is not seen and the area appears flattened.

It may be difficult to differentiate valvular and infundibular stenosis, and furthermore both types may be present. If the stenosis is infundibular rather than valvular, the thrill in the pulmonary artery is coarser and of lower frequency and is felt over a wider area, and a contraction wave can be seen

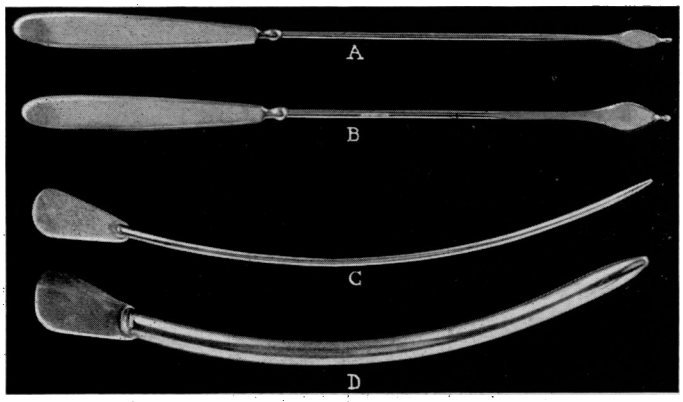


FIG. 7.—Pulmonary valvulotomes and sounds devised by Mr. Brock and Mr. Schrantz. (A) small valvulotome; (B) large valvulotome; (C) pulmonary artery probe; and (D) pulmonary artery sound.

proceeding from the infundibular chamber to the pulmonary artery. Beyond the infundibular chamber the pulmonary artery is not noticeably enlarged and the sinuses of Valsalva may be visible; also the jet effect is largely dissipated before the blood passes through the pulmonary valve. In doubtful cases pressure measurements in the right ventricle, in the pulmonary artery, and in a suspected infundibular chamber may be of help in differential diagnosis. We expect to utilize such measurements to a greater extent in the future.

If the diagnosis of valvular pulmonic stenosis is confirmed by inspection and palpation, 0.5 per cent procaine is injected into the myocardium of the right ventricle about 6 cm. caudad to the region of the pulmonary valve. Temporary irregularity of the heart may result, and a decline in blood pressure may occur, in which case one should proceed with valvulotomy as rapidly as possible. Two stay sutures are placed several centimeters apart in a longitudinal direction in the myocardium of the right ventricle at the site where the procaine had been injected (Fig. 6). An incision parallel to, and between

these sutures is made with a small sharp knife. This incision extends only partially through the ventricular wall. One may then use the instruments which were designed by Mr. Brock for location, division and dilatation of the stenotic valve. These are shown in Figure 7. The first is a curved probe with a small tip with which one may locate the cavity of the right ventricle and determine the angle at which the instrument must turn in order to enter the pulmonary artery. Furthermore, some estimate as to the degree of stenosis may be obtained by noting the resistance offered to the passage of the probe. After removal of the probe the small flat Brock valvulotome is introduced into the right ventricle with its broad surface parallel to the direc-

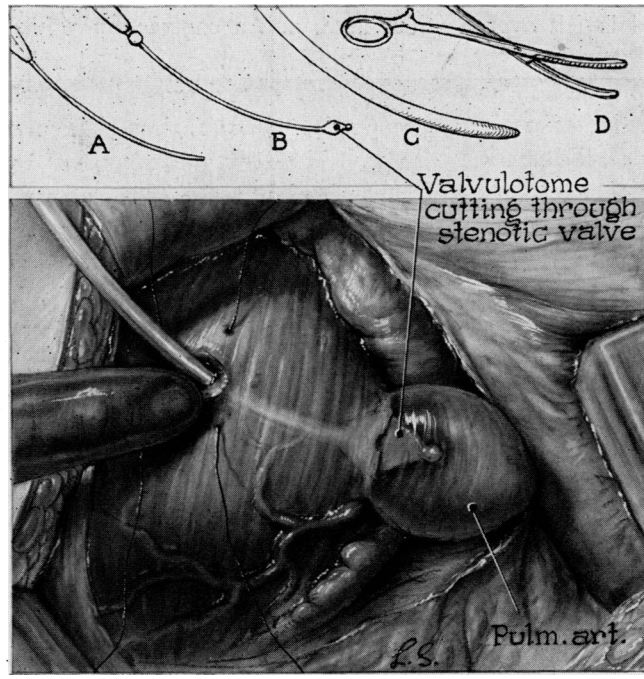


FIG. 8.—Transparency to demonstrate cutting of pulmonary valve with the flat Brock valvulotome.

tion of the incision (Fig. 8). The main secret of the success of this operation is this simple instrument.* The valvulotome has a gently curved shaft ending in a blade shaped like a spearhead with a short blunt probelike end. The two edges proximal to the probe-end are sharp and cutting and the shoulders and the retreating edges are blunt. This flat valvulotome is introduced through the incision in the right ventricle and through the stenotic valve until the probe-end can be palpated in the pulmonary artery. In most instances the resistance offered to the passage of the valvulotome is not great, since the walls

* Made by Mr. Schrantz, Genito-Urinary Manufacturing Company, 28a Devonshire Street, London, W.1, England.

of the stenotic valve are usually fairly thin and the blades of the valvulotome are sharp. Following removal of the instrument, bleeding is controlled by pressure with the index finger and slight traction on the stay sutures. A larger valvulotome is introduced, resulting in further division of the stenotic valve. At present we have a choice of valvulotomes of three sizes and propose to have others made of varying widths.

After a large cutting valvulotome has been used, a circular sound is passed with which the valve is dilated (Fig. 9). The passage of one or more sounds is followed by the introduction of a curved clamp which is opened partially at the proper place, resulting in further stretching and tearing of the valve.

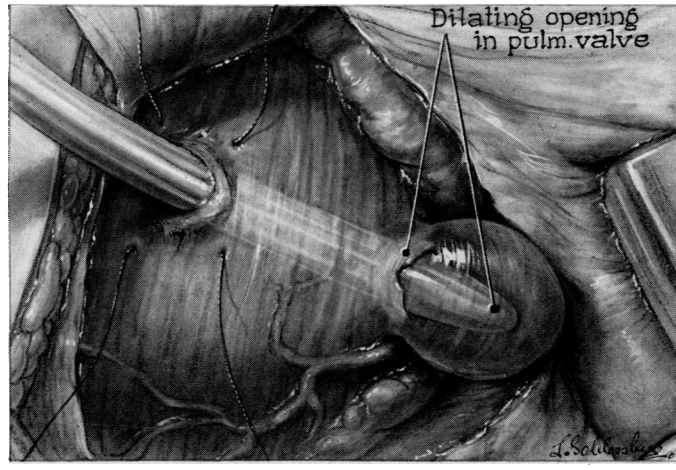


FIG. 9.—Transparency to demonstrate dilatation of previously divided pulmonary valve by passage of curved sound.

None of the valve has been removed in the operations herein reported, although it would be feasible to do so if it were considered advisable. After the desired degree of cutting and dilatation of the valve has been attained, the incision in the myocardium is closed with interrupted sutures and the stay sutures are removed. The action of the heart frequently becomes poor while the instrumentation is being carried out and the arterial pressure declines. Improvement, however, usually takes place promptly after manipulation is stopped. Palpation of the pulmonary artery then reveals a lower pitched thrill, evidence of increased pressure, and a prominence in the region of the sinuses of Valsalva which was not present before. The pericardium is loosely closed and a catheter is placed in the pleural cavity through an intercostal space. The incision in the chest wall is closed in multiple layers with interrupted sutures. The entire operating time on children is only about an hour and most of this is spent in making and closing the incision in the chest.

We wish to emphasize a point made by Mr. Brock, which is that the action of the heart may become very poor shortly after this organ is exposed.

A decline in the systemic arterial blood pressure in the presence of severe pulmonic stenosis results in definite dimunition in the flow of blood to the lungs, and the entire circulation, including that to the myocardium, suffers. Under such circumstances one should proceed immediately with division of the stenotic valve rather than wait with the hope that the action of the heart will improve spontaneously. Recently, Drinalfa has been injected intramuscularly just before the heart is exposed, and it is our impression that it has been helpful in preventing a fall in blood pressure.

PRESENTATION OF CLINICAL MATERIAL

The material in this report consists of 19 patients, all of whom underwent pulmonary valvulotomy by the Brock technic between November 1, 1949, and March 15, 1950. These patients ranged in age from 14 months to 20 years, the average age being eight years. There were 12 females and seven males. Only two of the patients gave no history of cyanosis. Five patients were noted to be cyanotic at birth, and in the remaining 12 cyanosis developed at some time between six months and five years of age. As can be seen in the table, ten of the 17 patients in whom the peripheral arterial oxygen saturation was determined preoperatively showed less than 80 per cent saturation. One of the patients in whom this determination unfortunately was not made was intensely cyanotic.

As regards exercise tolerance, all but one of these patients (M. K.) showed great limitation of activity. Most of them were able to walk one to four city blocks on level ground without resting, but three were unable to walk more than a few feet. Only five of these patients gave a history of squatting, whereas in patients with the tetralogy of Fallot this is an almost universal finding. In 17 of the patients the liver edge could be felt 1 to 6 cm. below the right costal margin and in 16 of them definite pulsations could be detected. In the one instance where there was no hepatomegaly it was our impression that the interauricular septal defect was unusually large.

All but one patient had a loud, harsh systolic murmur in the pulmonary area at the time of operation. The patient in whom no murmur was audible (A. W.) had had one when seen three months previously. Associated with the disappearance of the murmur was a deterioration in the patient's condition and a deepening of the cyanosis. At operation the findings indicated that the opening in the pulmonary valve had closed completely and that the patient's life was sustained by collateral circulation. Except in two instances, cardiac enlargement was present, and 14 patients had a cardio thoracic ratio of 60 per cent or above. In almost all instances this cardiomegaly was associated with prominence of the precordium.

As can be seen in the table, a number of these patients had some degree of polycythemia, and the increase in the number of red cells was, in a rough way, inversely proportional to the arterial oxygen saturation. On reference

to the table it can be seen that in all instances where cardiac catheterization was successfully carried out there was a significant degree of right ventricular hypertension. It is interesting that the girl (M. K.) who was found to have the highest pressure in the right ventricle (223/13 mm. mercury) was the only patient in whom there was no serious impairment in exercise tolerance. She was able to carry on a normal but quiet way of life and could walk about one mile on level ground. Radiologic studies in these patients consistently revealed diminished vascularity of the lung fields, right-sided enlargement of the heart, and marked prominence of the main pulmonary artery. The electrocardiogram in two instances demonstrated a right bundle branch block. The other tests showed right axis deviation with patterns suggestive of right auricular and right ventricular hypertrophy.

It is a tribute to Doctor Taussig and her group that in each case the preoperative diagnosis proved to be correct. Three of the patients, however, had previously been thought to have the tetralogy of Fallot and had had an artificial ductus created. Two of these patients had been operated on in The Johns Hopkins Hospital more than two years previously and had been in severe right-sided heart failure since shortly after the operation. In the other patient an anastomosis had been performed in another hospital six weeks prior to valvulotomy and prompt and severe failure had developed.

During operation all these patients demonstrated a rather constant pattern of behavior. Until the valve was divided they were in a precarious state, when, as has been emphasized by Brock, any undue delay while awaiting improvement in the patient's condition might well have proved fatal. There was sometimes a transient decline in blood pressure during anesthesia; in each case the systolic pressure fell 20 to 60 mm. of mercury at the time the pericardium was opened and in four instances the pressure became inaudible. However, with expeditious division of the valve, the heart action usually improved promptly and the pressure returned to the preoperative levels. This fact was most strikingly demonstrated in the patient who had no murmur at the time of operation (A. W.). She was intensely cyanotic from the outset of the operation, and by the time the pericardium was incised her blood pressure was unobtainable and the heart action was very weak. By the time the first instrument was passed the contractions were scarcely perceptible. Within one minute following division of the valve the heart showed vigorous, forceful contractions and the blood was a normal red. Another patient survived two brief periods of cardiac arrest. Aside from extrasystoles caused by manipulation of the heart, no arrhythmias were noted.

Postoperatively the patients were kept in an oxygen tent for about 48 hours and careful attention was paid to fluid balance. In patients who had been digitalized preoperatively, digitalis was continued for a week or two after operation and then discontinued. One patient (E. W.) who had been in severe failure following creation of an artificial ductus two years previously and who required a second procedure to close the ductus, was discharged

from the hospital on a maintenance dose of digitalis. All patients were given prophylactic doses of penicillin for ten days postoperatively. It is interesting that for five to ten days after operation these patients had an elevation of temperature of greater degree than one might expect. It was thought that this might have been related to the myocardial damage produced by the incision in the right ventricle. The patients were kept in the hospital for three weeks and were allowed to get out of bed during the last week.

There have been two deaths in this series of 19 cases. The first occurred in the first operation in this series in a 14-year-old boy (H. W.) who had been operated on about two and one-half years previously with the diagnosis of tetralogy of Fallot; an artificial ductus had been created on the left side. About two weeks after operation signs of right-sided heart failure appeared. Since then he had been maintained on digitalis and had been virtually confined to bed. He was in very poor condition with extreme cardiac enlargement, a large pulsating liver, auricular fibrillation, peripheral edema, and three plus albuminuria. Except for a severe fall in blood pressure he tolerated valvulotomy fairly well. It was then felt advisable to attempt closure of the artificial ductus. Shortly after this dissection was undertaken his heart action became very weak and finally ceased. It was revived with adrenalin and massage but could not be maintained, and the patient died on the operating table. At autopsy the pulmonary valve was found to have been divided. Since then there have been two other patients who previously had creation of an artificial ductus. In one of these patients the anastomosis was successfully closed at the time of valvulotomy and in the other the anastomosis was closed two weeks subsequent to the valvulotomy. The latter of these two patients remained in severe failure with a large pulsating liver after valvulotomy, but made rapid improvement after closure of the anastomosis.

The second death occurred in a 9-year-old girl (J. T.), the eighth patient in this series, who had a severe degree of stenosis accompanied by intense cyanosis and right ventricular hypertension (see Table I). She tolerated the operation well and immediately her color was excellent and the liver pulsations were strikingly diminished. Forty-eight hours after operation following several hours of restlessness and mental confusion she suddenly became comatose and voluntary respiratory effort ceased. Artificial respiration was maintained through an endotracheal airway, and the patient was again taken to the operating room where a trephine was carried out by Dr. Robert Fisher and a left frontal lobe abscess was drained. After this procedure the child was kept in a respirator, but pursued a progressively downhill course, dying about 72 hours after the valvulotomy. Autopsy again demonstrated successful division of the pulmonary valve as well as a chronic brain abscess of the left frontal lobe. For several weeks before coming to the hospital the child had had mild recurrent headaches and on admission there was leukocytosis (15,000 wbc/c.mm.). The significance of these symptoms, however, was not appreciated, and the presence of a brain abscess was not suspected. Except

for this previously existing abscess, she would almost certainly have been greatly benefited.

The other 17 patients are alive and in good condition. It is gratifying that there has been no serious postoperative complication in these cases. All the patients who were cyanotic preoperatively showed an improvement in color immediately following valvulotomy, and in several patients who had been intensely cyanotic the color promptly became normal. It is interesting that on the second or third postoperative day, particularly during exertion such as coughing or crying, the cyanotic hue sometimes re-appeared, usually to a lesser degree than before, and then subsided slowly over a period of several

TABLE I.—*Data Relative to Patients Discussed in Text.*

Patient		Age—Years	Cardio-thoracic Ratio Per cent	Red Cell Count millions/c.mm.	Right Ventricular Pressure mm./mercury	Preoperative Arterial Oxygen Saturation Per cent	Postoperative Arterial Oxygen Saturation Per cent
H.W.	♂	14	64	8.4	.....	72.8	Deceased
C.S.	♀	7	70	6.9	162/45	68.0	95.7
A.V.	♂	7	72	8.8	125/0	70.0	94.5
R.F.	♀	5	45	7.4	98/?	62.0	88.0
C.S.	♂	14	40	5.1	122/30	87.0	97.3
E.W.	♂	9	63	5.2	.....	82.1	....
T.S.	♂	1-2/12	..	8.9	85/?	....	....
J.T.	♀	9	62	7.3	191/30	76.0	Deceased
R.H.	♂	20	70	9.0	150+/?	68.0	91.4
C.B.	♂	13	62	5.5	143/0	95.6	92.4
E.F.	♀	4	57	5.3	165/36	92.3	94.5
A.W.	♀	2	63	11.8	.....	....	85.0
E.K.	♀	6	65	6.5	146/48	65.8	88.0
E.B.	♀	2	60	6.6	.....	66.0	82.0
M.K.	♀	13	60	5.2	223/13	93.5	89.4
C.J.	♀	11	60	6.3	.....	85.0	92.3
E.S.	♀	7	64	5.8	166/26	83.6	94.0
S.L.	♀	1-3/12	70	5.6	84/60	71.0	88.7
R.B.	♀	7	53	5.5	200/?	78.0	85.5

hours. As can be seen in the table a number of the patients showed a definite rise in the arterial oxygen saturation and in several this became normal.

Another striking change which was detected immediately after operation was the alteration in the character of the murmur. The original loud, high-pitched systolic murmur was usually converted into a low-pitched rumble which at times was very soft and no longer accompanied by a thrill. Liver pulsations also promptly diminished or disappeared. Only two patients at the time of discharge were believed to have faint pulsations in the liver, whereas all but one had demonstrated this sign preoperatively. After valvulotomy the size of the liver fluctuated considerably from day to day, but the general trend was a slow diminution in size. In two patients during the early postoperative period the liver edge was felt to be lower than it was preoperatively, but by the time of discharge the liver was smaller than it had been on admission. Where there had been polycythemia there was a return to more nearly normal

levels, but the validity of this observation is open to question because of the factor of blood loss at operation. Ten patients had had albuminuria on admission but this condition had cleared in nine by the time they left the hospital. The electrocardiogram showed no significant change during the observation period.

As regards exercise tolerance, it is difficult to assess improvement accurately because all of the patients are still on restricted activity. However, five of the patients had shown a high degree of exercise intolerance, and even during the period of hospitalization it was obvious that their capacity for physical activity had improved satisfactorily.

Four of these patients have recently returned to the Cardiac Clinic of the Harriet Lane Home for examination four months after operation. One of them (C. S.♀) had gained 21 pounds in weight and tolerated all allowed exercise without any sign of dyspnea or fatigue. Her red blood count had dropped about one million from the preoperative level and her cardiothoracic ratio had declined from 70 per cent to 60 per cent. Another child (A. V.) likewise had an excellent color and his mother was having difficulty in restraining his activity. His red blood count had fallen from 8.8 million per c.mm. to 4.7 million and his cardiothoracic ratio had fallen from 72 per cent to 61 per cent. In the other two patients the results were less striking because their original disability had been less severe. Both of them had a good color, but one of them (R. F.) is said still to have occasional periods of cyanosis though this was not apparent at the clinic. Neither had been active enough to determine his exercise tolerance, though one (C. S.♂) had walked as far as one mile, having been able to walk only one-half block preoperatively. These patients were placed on unrestricted activity except for C. S.♂, in whom the heart sounds were not of good quality although there was no other evidence of cardiac difficulty. He was advised to remain on restricted activity for another three-month period.

The most objective method for determining improvement would be to repeat cardiac catheterization in those patients for whom we have preoperative data. It is felt that it is still too soon after operation to carry out these studies, but they are planned for the future.

DISCUSSION

Technically this operation is not a difficult procedure. Its success depends on careful attention to detail from the beginning of the diagnostic work-up until the patient has passed through the immediate postoperative period. Much responsibility rests upon the anesthetist, who should observe the condition of the patient with particular care as soon as the chest has been opened. There may be nothing specific in the various medications which are administered both before and during operation, but we feel that each drug in its proper place fulfills a definite function.

One essential which should be stressed is adequate exposure, and this can usually be obtained by the left anterolateral approach described above, especially if the heart is enlarged. This exposure is necessary not only for instrumentation but also for adequate assessment of the situation by direct vision and palpation. While it is true that the typical case can be accurately diagnosed by preoperative studies, there will probably be many cases where the mode of treatment will depend to a large degree on operative findings. It is of the utmost importance that the various valvulotomes and probes be passed well into the pulmonary artery, because it is possible to push the diaphragmatic valve in front of the instrument for a short distance into the artery without actually dividing the valve. Usually one is aware of a sudden release in resistance as the valve gives way, but it is well to check the position of the instrument by palpating the artery beyond the valve. Also it should be borne in mind that the stay sutures function only as guides and should not be subjected to strong traction lest they be torn loose. Bleeding is best controlled by pressure with the finger. Blood loss is not likely to be great if the ventricular wall is very hypertrophic. If, however, the myocardium has been thinned out by dilatation, hemorrhage may be troublesome and require rapid replacement of blood.

One of the most interesting anatomical features of these cases, as has been commented upon above, has been the regular occurrence of marked dilatation of the main pulmonary artery just beyond the stenotic valve. At times this has attained impressive proportions and occasionally the artery has been three or four times greater in diameter than normal. This is an important diagnostic sign both radiologically and at the time of operation when the artery is exposed. Such dilatation of an artery beyond an obstruction is a well-known phenomenon which has been recognized for many years, particularly where it occurs in the subclavian artery distal to a cervical rib or in the aorta below a coarctation. Doctor Halsted[5] studied this problem in experimentally produced constrictions of the aorta and found that a considerable degree of stenosis was necessary to produce a significant dilatation of the distal portion of the vessel. He also noted an elevation of diastolic pressure and lowering of systolic pressure beyond the constriction. He felt that this lowering of the pulse pressure and the abnormal play of the blood stream produced by the powerful jet were the chief mechanical factors responsible for the dilatation. In our cases where the dilatation was very great it will be of interest to see what the ultimate fate of the artery will be. Once the valve has been divided the jet effect is largely abolished, but the artery appears even larger once it is subjected to increased intraluminal pressure. So far there has been no indication that this vessel will not function efficiently after valvulotomy.

A number of important problems present themselves in regard to this operation, some of which cannot be answered with assurance at the present time. One of the most pressing of these problems is the ultimate fate of the

incised valve. Is there any danger that the cut edges will heal together, or will the contraction of scar tissue lead to the reformation of a constriction? Also it is pertinent to ask whether or not this procedure creates an incompetent pulmonary valve which might eventually result in a serious degree of pulmonary insufficiency. It must be remembered that this operation does not merely produce a dilatation of the stenotic valve but is designed to actually divide the valve through its transverse axis from one side of the ring to the other. In such a clean surgical wound one would expect endothelialization of the cut edges to take place within a few days. On the basis of a four-month period of postoperative observation we can say only that thus far there has been no evidence of reformation of the obstruction. It is possible in the future it will be found that mere division of the valve is not adequate and that it will be desirable to actually remove a portion of the valve. Such a defect would be expected to produce some degree of pulmonary insufficiency, but in the absence of pulmonary disease producing pulmonary hypertension it does not seem likely that the valvular incompetence would be attended by dangerous consequences. Powers and Bowie[6] in 1933 excised segments of the pulmonary valve in two dogs in which a pulmonary stenosis had previously been experimentally produced. Diastolic murmurs developed but the dogs survived for 20 months without sign of decompensation. In our series of cases there has been no auscultatory evidence of pulmonary insufficiency. Moreover, in the autopsy specimens of the two patients who died the divided diaphragm looked as though it might well function as a reasonably competent bicuspid valve.

Another question which comes to mind is the fate of the interauricular septal defect which is an associated finding in most of these cases. At the present time there is not enough available information from autopsies to speak accurately about this point. It is believed, however, largely on the basis of catheterization data, that in most instances the shunt takes place through the foramen ovale which has been held open preoperatively by the abnormal pressure gradient between the two auricles. The surgical correction of the obstruction at the pulmonary valve undoubtedly reduces this pressure gradient and in some cases may bring about either functional or anatomical closure of the foramen. In some instances the defect is undoubtedly too large to allow spontaneous closure. These patients (such as R. F.) may continue to have brief periods of cyanosis when engaged in strenuous exercise. Also it is possible that there might eventually be a reversal of the shunt through the defect, the flow being from left to right as is the case when an interauricular septal defect occurs as an isolated anomaly. Such a situation would place an additional burden on the right side of the heart.

One also wonders about the effect of pulmonary valvulotomy on the size of the enlarged heart. It is the opinion of Doctor Taussig[7] that the amount of enlargement of the cardiac silhouette which is due to right ventricular dilatation can be expected to decrease promptly, but she looks upon the

myocardial hypertrophy as a more or less permanent feature. When pulmonary stenosis is relieved at an early age, the child may "grow into the size of his heart" and the cardiothoracic ratio will become more nearly normal. In several of the patients in our series a significant decline in the cardiothoracic ratio was noted four months postoperatively. It is likely that much of this reduction has taken place in the right auricular salient of the cardiac silhouette.

As has been shown by Sellers and by Brock and, as we believe, has been demonstrated by this series of cases, surgical division of the congenitally stenotic pulmonary valve is a technically feasible procedure. The mortality rate is not prohibitive when one considers the poor prognosis of the patient in whom a severe degree of stenosis exists. But when one is asked to evaluate such a patient for operation, he is faced with serious problems. What is the optimal age for such an operation? Will the opening which is created in the pulmonary valve of a young child still be adequate when the patient is full grown? Is there any basis for expecting that this orifice will enlarge as the child develops? So far this decision has not been difficult to make because most of our patients have been in such precarious condition that a prolonged delay in operation was believed to be unwarranted. Also, of course, it is desirable that the obstruction be relieved at an early age before severe and perhaps irreversible damage has been done to the myocardium, pulmonary artery, liver, and kidneys. Again we must wait on time and careful follow-up studies to solve these problems.

Congenital valvular pulmonic stenosis with intact ventricular septal defect is of less frequent incidence than our recent experience would indicate. Some of these patients had been seen previously in the Cardiac Clinic and operation had been postponed. It is Doctor Taussig's present impression that the incidence of valvular pulmonic stenosis is about one-tenth as great as that of the tetralogy of Fallot.

SUMMARY

Division of the stenotic pulmonary valve by the method of Brock has been performed on 19 patients in The Johns Hopkins Hospital in the treatment of congenital valvular pulmonic stenosis with intact ventricular septum. There have been two deaths, one attributable to severe heart failure and one due to a pre-existing abscess of the brain. The remaining patients are improved and some appear to be in normal health. There have been no deaths during or following the last 11 operations. Valvular pulmonic stenosis is a mechanical disorder which can be diagnosed by available methods of study and which can be treated by direct operative attack on the valve.

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DISCUSSION.—DR. WILLIS J. POTTS: That was a very fascinating presentation of a new method for approaching a certain type of patient with congenital heart disease.

The so-called pure pulmonary stenosis often is not a single lesion, as the name implies, but is associated with an atrial septal defect. This condition accounts for a small but important group of cyanotic or very slightly cyanotic children. The degree of cyanosis is dependent upon the size of the inter-auricular defect. It is obvious, as Dr. Blalock has pointed out, that a shunt operation is contraindicated in such patients. Brock's approach is entirely logical.

[Slide] This demonstrates—as did Dr. Blalock's picture—the typical picture of so-called pure pulmonary stenosis. Looking in the pulmonary artery toward the heart, one sees the conelike constriction of the pulmonary valves. The stenotic structure projecting into the pulmonary artery resembles an infantile cervix extending into the vagina.

[Slide] In preparation for operation on such a patient, we devised a knife to open this constriction. The blade of the best cataract knife available was soldered to a rounded shaft of such size that it will fill the hole made in the ventricle by the blade. The snug fit of this shaft in the heart muscle prevents loss of blood. The rest of the handle of the knife is standard, for secure handling.

Two patients with isolated pulmonary stenosis have been operated upon with this instrument. The first child, 23 days old, weighed seven pounds, and was operated upon in December, 1949. The infant was mildly cyanotic, had some enlargement of the heart, and could not live outside an oxygen tent. From our experimental work on dogs, we planned an approach to the stricture through the pulmonary artery, but at operation on this child, the transventricular approach seemed more feasible.

[Slide] The heart was exposed through a curved submammary incision. The heart was lifted from the pericardial sac, and two holding sutures were put in the wall of the right ventricle, relatively near the base of the heart. The knife was then thrust into the right ventricle and directed toward the pulmonary artery, with the sharp edge of the blade directed toward the lumen. The constriction was cut by three radial incisions from the periphery inward. The knife was withdrawn and the wound in the ventricle closed with one stitch. The child made a rather spectacular recovery, and is doing well today, after four months.

The second child was 14 months old, moderately cyanotic, and subject to frequent attacks of unconsciousness. A similar operation was done on this child about two weeks ago. Immediately after the constriction was cut, pressure in the pulmonary artery was palpably higher. The child's color improved on the operating table, and has remained good thus far—only two weeks.

As you see in the illustration, there was some constriction of the walls of the pulmonary artery where it emerges from the heart.

The knife described worked well in these two infants. In older children with this deformity, Brock's double-bladed knife or some modification of it would be better.